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## SEQUENCE LISTING

<110> Egelrud, Torbjorn  
Hansson, Lennart

<120> SCCE modified transgenic mammals and  
their use as models of human diseases

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 <212> PRT  
 <213> Artificial Sequence

<220>  
 <223> Consensus sequence for cleavage site in C-terminal  
 of SCCE.

<221> VARIANT  
 <222> 2  
 <223> Asp = either aspartate (Asp) or glutamate (Glu).

<221> VARIANT  
 <222> 3  
 <223> Lys = either lysine (Lys) or arginine (Arg).

<400> 14  
 Gly Asp Lys Ile Ile Asp Gly  
 1 5

<210> 15  
 <211> 8  
 <212> PRT  
 <213> Artificial Sequence

<220>  
 <223> consensus of the substrate specificity pouch.

<221> VARIANT  
 <222> 1  
 <223> Thr = any amino acid residue.

<221> VARIANT  
 <222> 3  
 <223> Ala = any amino acid residue.

<221> VARIANT  
 <222> 5  
 <223> Asn = any amino acid residue.

<400> 15  
 Thr Asn Ala Cys Asn Gly Asp Ser  
 1 5



9

<210> 16  
<211> 20  
<212> DNA  
<213> Artificial sequence

<220>  
<223> PCR primer SYM3300.

<400> 16  
ggtggccctg ctcagtggca 20

<210> 17  
<211> 22  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM3301.

<400> 17  
caccatggat gacacagcct gg 22

<210> 18  
<211> 20  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM3302.

<400> 18  
aataaagaaa cacaaaaccc 20

<210> 19  
<211> 18  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM3418.

<400> 19  
tgtaatatca ttgtgggc 18

<210> 20  
<211> 18  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM4118.

<400> 20  
ggatgtgaag ctcattctc 18

<210> 21  
<211> 18

10

<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM4121.

<400> 21  
tggagtcggg gatgccag

18

<210> 22  
<211> 25  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM4720.

<400> 22  
gggaggggtgg agagagagtg cagtg

25

<210> 23  
<211> 21  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer SYM4899.

<400> 23  
agtctaggct gcagccccta c

21

<210> 24  
<211> 22  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer hEXON1.

<400> 24  
ctcgagggat ctgatgtgat cc

22

<210> 25  
<211> 24  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer mEXON1.

<400> 25  
ctgggagtga cttggcgtgg ctct

24

<210> 26  
<211> 23  
<212> DNA

<213> Artificial Sequence

<220>

<223> PCR primer specific for human SCCE IE2.

<400> 26  
gctctcccat tagtccccag aga 23

<210> 27  
<211> 24  
<212> DNA  
<213> Artificial Sequence

<220>

<223> PCR primer specific for human SCCE MJ2.

<400> 27  
ccacttggtg aacttgacac cttg 24

<210> 28  
<211> 18  
<212> DNA  
<213> Artificial Sequence

<220>

<223> forward primer covering the position 427 - 444 of the human SCCE cDNA sequence.

<400> 28  
gggaaccccc tggaacaa 18

<210> 29  
<211> 21  
<212> DNA  
<213> Artificial Sequence

<220>

<223> reverse primer covering the position 490 - 510 of the human cDNA sequence in exon five.

<400> 29  
acatccacgc acatgaggtc a 21

<210> 30  
<211> 29  
<212> DNA  
<213> Artificial Sequence

<220>

<223> The real time amplification probe covering the position 445 - 473 of the human cDNA sequence in exon four.

<400> 30  
cctgtactgt ctccggctgg ggcactacc 29

12

<210> 31  
<211> 25  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer mS3.

<400> 31  
caaggagaaa ggattataga tggct

25

<210> 32  
<211> 21  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer 698.

<400> 32  
aaggctccgc acccatggca g

21

<210> 33  
<211> 26  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer 696.

<400> 33  
tgcaatggtg actcaggggg gccott

26

<210> 34  
<211> 23  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer H2.

<400> 34  
gacccaggcg tctacactca agt

23

<210> 35  
<211> 25  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer mS4.

<400> 35  
gagaccatga aaacccatcg ctaac

25

<210> 36

13

<211> 26  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer KO 0905.

<400> 36  
tgactttctt cacactggac gacagc

26

<210> 37  
<211> 26  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer GR 0905.

<400> 37  
cttcacactg gctgatagcc tggccg

26

<210> 38  
<211> 28  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer Ngr.

<400> 38  
cagggtggcg gaatgacctc atggccct

28

<210> 39  
<211> 28  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer RA 1016.

<400> 39  
ctactccaca aggacccatg tcaatgac

28

<210> 40  
<211> 28  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> PCR primer nRA 1016.

<400> 40  
gctgtgtgct ggcattcccg actctaag

28

<210> 41  
<211> 30

14

<212> DNA  
<213> Artificial Sequence

<220>  
<223> SMART II oligonucleotide.

<400> 41  
aagcagtggg aacaacgcag agtacgcggg 30

<210> 42  
<211> 27  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> 5'-RACE cDNA synthesis primer.

<221> variation  
<222> 27  
<223> n = a or g or c or t

<400> 42  
tttttttttt tttttttttt tttttvn 27

<210> 43  
<211> 45  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> Long universal primer.

<400> 43  
ctaatacgac tcactatagg gcaagcagtg gtaacaacgc agagt 45

<210> 44  
<211> 23  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> Short universal primer.

<400> 44  
ctaatacgac tcactatagg gcc 23

<210> 45  
<211> 23  
<212> DNA  
<213> Artificial Sequence

<220>  
<223> Nested universal primer.

<400> 45  
aagcagtggg aacaacgcag agt 23

<210> 46  
<211> 243  
<212> PRT

15

&lt;213&gt; Artificial Sequence

&lt;220&gt;

<223> Deduced amino acid sequence from the C-terminal  
part of SCCE from cow.

&lt;400&gt; 46

```

Met Thr Thr Pro Leu Val Ile Leu Leu Leu Thr Phe Ala Leu Gly Ser
 1          5          10          15
Val Ala Gln Glu Asp Gln Gly Asn Lys Ser Gly Glu Lys Ile Ile Asp
          20          25          30
Gly Val Pro Cys Pro Arg Gly Ser Gln Pro Trp Gln Val Ala Leu Leu
          35          40          45
Lys Gly Ser Gln Leu His Cys Gly Gly Val Leu Leu Asn Glu Gln Trp
          50          55          60
Val Leu Thr Ala Ala His Cys Met Asn Glu Tyr Asn Val His Met Gly
          65          70          75          80
Ser Val Arg Leu Val Gly Gly Gln Lys Ile Lys Ala Thr Arg Ser Phe
          85          90          95
Arg His Pro Gly Tyr Ser Thr Gln Thr His Ala Asn Asp Leu Met Leu
          100          105          110
Val Lys Leu Asn Gly Arg Ala Lys Leu Ser Ser Ser Val Lys Lys Val
          115          120          125
Asn Leu Pro Ser His Cys Asp Pro Pro Gly Thr Met Cys Thr Val Ser
          130          135          140
Gly Trp Gly Thr Thr Thr Ser Pro Asp Val Thr Phe Pro Gly Gln Leu
          145          150          155          160
Met Cys Thr Asp Val Lys Leu Ile Ser Pro Gln Asp Cys Arg Lys Val
          165          170          175
Tyr Lys Asp Leu Leu Gly Asp Ser Met Leu Cys Ala Gly Ile Pro Asn
          180          185          190
Ser Arg Thr Asn Ala Cys Asn Gly Asp Ser Gly Gly Pro Leu Met Cys
          195          200          205
Lys Gly Thr Leu Gln Gly Val Val Ser Trp Gly Ser Phe Pro Cys Gly
          210          215          220
Gln Pro Asn Asp Pro Gly Val Tyr Thr Gln Val Cys Lys Tyr Val Asn
          225          230          235          240
Trp Ile Lys

```

&lt;210&gt; 47

&lt;211&gt; 249

&lt;212&gt; PRT

&lt;213&gt; Artificial Sequence

&lt;220&gt;

<223> Deduced amino acid sequence from the C-terminal  
part of SCCE from pig.

&lt;400&gt; 47

```

Met Ala Arg Pro Leu Leu Pro Pro Arg Leu Ile Leu Leu Leu Ser Leu
 1          5          10          15
Ala Leu Gly Ser Ala Ala Gln Glu Gly Gln Asp Lys Ser Gly Glu Lys
          20          25          30
Ile Ile Asp Gly Val Pro Cys Pro Gly Gly Ser Arg Pro Trp Gln Val
          35          40          45
Ala Leu Leu Lys Gly Asn Gln Leu His Cys Gly Gly Val Leu Val Asn
          50          55          60
Gln Gln Trp Val Leu Thr Ala Ala His Cys Met Met Asn Asp Tyr Asn
          65          70          75          80

```

16

Val His Leu Gly Ser Asp Arg Leu Asp Asp Arg Lys Gly Gln Lys Ile  
 85 90 95  
 Arg Ala Met Arg Ser Phe Arg His Pro Gly Tyr Ser Thr Gln Thr His  
 100 105 110  
 Val Asn Asp Leu Met Leu Val Lys Leu Ser Arg Pro Ala Arg Leu Ser  
 115 120 125  
 Ala Ser Val Lys Lys Val Asn Leu Pro Ser Arg Cys Glu Pro Pro Gly  
 130 135 140  
 Thr Thr Cys Thr Val Ser Gly Trp Gly Thr Thr Thr Ser Pro Asp Val  
 145 150 155 160  
 Thr Phe Pro Ala Asp Leu Met Cys Thr Asp Val Lys Leu Ile Ser Ser  
 165 170 175  
 Gln Asp Cys Lys Lys Val Tyr Lys Asp Leu Leu Gly Ser Ser Met Leu  
 180 185 190  
 Cys Ala Gly Ile Pro Asn Ser Lys Thr Asn Ala Cys Asn Gly Asp Ser  
 195 200 205  
 Gly Gly Pro Leu Val Cys Lys Gly Thr Leu Gln Gly Leu Val Ser Trp  
 210 215 220  
 Gly Thr Phe Pro Cys Gly Gln Pro Asn Asp Pro Gly Val Tyr Thr Gln  
 225 230 235 240  
 Val Cys Lys Tyr Ile Asp Trp Ile Asn  
 245

&lt;210&gt; 48

&lt;211&gt; 253

&lt;212&gt; PRT

&lt;213&gt; Artificial Sequence

&lt;220&gt;

<223> Deduced amino acid sequence from the C-terminal  
 part of SCCE from homo.

&lt;400&gt; 48

Met Ala Arg Ser Leu Leu Pro Leu Gln Ile Leu Leu Leu Ser Leu  
 1 5 10 15  
 Ala Leu Glu Thr Ala Gly Glu Glu Ala Gln Gly Asp Lys Ile Ile Asp  
 20 25 30  
 Gly Ala Pro Cys Ala Arg Gly Ser His Pro Trp Gln Val Ala Leu Leu  
 35 40 45  
 Ser Gly Asn Gln Leu His Cys Gly Gly Val Leu Val Asn Glu Arg Trp  
 50 55 60  
 Val Leu Thr Ala Ala His Cys Lys Met Asn Glu Tyr Thr Val His Leu  
 65 70 75 80  
 Gly Ser Asp Thr Leu Gly Asp Arg Arg Ala Gln Arg Ile Lys Ala Ser  
 85 90 95  
 Lys Ser Phe Arg His Pro Gly Tyr Ser Thr Gln Thr His Val Asn Asp  
 100 105 110  
 Leu Met Leu Val Lys Leu Asn Ser Gln Ala Arg Leu Ser Ser Met Val  
 115 120 125  
 Lys Lys Val Arg Leu Pro Ser Arg Cys Glu Pro Pro Gly Thr Thr Cys  
 130 135 140  
 Thr Val Ser Gly Trp Gly Thr Thr Thr Ser Pro Asp Val Thr Phe Pro  
 145 150 155 160  
 Ser Asp Leu Met Cys Val Asp Val Lys Leu Ile Ser Pro Gln Asp Cys  
 165 170 175  
 Thr Lys Val Tyr Lys Asp Leu Leu Glu Asn Ser Met Leu Cys Ala Gly  
 180 185 190  
 Ile Pro Asp Ser Lys Lys Asn Ala Cys Asn Gly Asp Ser Gly Gly Pro  
 195 200 205  
 Leu Val Cys Arg Gly Thr Leu Gln Gly Leu Val Ser Trp Gly Thr Phe



17

210		215		220
Pro Cys Gly Gln Pro Asn Asp Pro Gly Val Tyr Thr Gln Val Cys Lys				
225		230		235
Phe Thr Lys Trp Ile Asn Asp Thr Met Lys Lys His Arg				240
	245		250	

<210> 49  
 <211> 226  
 <212> PRT  
 <213> Artificial Sequence

<220>  
 <223> Deduced amino acid sequence from the C-terminal  
 part of SCCE from rat.

<400> 49  
 Met Gly Val Trp Leu Leu Ser Leu Leu Thr Val Leu Leu Ser Leu Ala  
 1 5 10 15  
 Leu Glu Thr Ala Gly Gln Gly Glu Arg Ile Ile Asp Gly Tyr Lys Cys  
 20 25 30  
 Lys Glu Gly Ser His Pro Trp Gln Val Ala Leu Leu Lys Gly Asp Gln  
 35 40 45  
 Leu His Cys Gly Gly Val Leu Val Gly Glu Ser Trp Val Leu Thr Ala  
 50 55 60  
 Ala His Cys Lys Met Gly Gln Tyr Thr Val His Leu Gly Ser Asp Lys  
 65 70 75 80  
 Ile Glu Asp Gln Ser Ala Gln Arg Ile Lys Ala Ser Arg Ser Phe Arg  
 85 90 95  
 His Pro Gly Tyr Ser Thr Arg Thr His Val Asn Asp Ile Met Leu Val  
 100 105 110  
 Lys Met Asp Lys Pro Val Lys Met Ser Asp Lys Val Gln Lys Val Lys  
 115 120 125  
 Leu Pro Asp His Cys Glu Pro Pro Gly Thr Leu Cys Thr Val Ser Gly  
 130 135 140  
 Trp Gly Thr Thr Thr Ser Pro Asp Val Thr Phe Pro Ser Asp Leu Met  
 145 150 155 160  
 Cys Ser Asp Val Lys Leu Ile Ser Ser Gln Glu Cys Lys Lys Val Tyr  
 165 170 175  
 Lys Asp Leu Leu Gly Lys Thr Met Leu Cys Ala Gly Ile Pro Asp Ser  
 180 185 190  
 Lys Thr Asn Thr Cys Asn Gly Asp Ser Gly Gly Pro Leu Val Cys Asn  
 195 200 205  
 Asp Thr Leu Gln Gly Leu Val Ser Trp Gly Thr Tyr Pro Cys Gly Gln  
 210 215 220  
 Pro Asn  
 225

<210> 50  
 <211> 249  
 <212> PRT  
 <213> Artificial Sequence

<220>  
 <223> Deduced amino acid sequence from the C-terminal  
 part of SCCE from mouse.

<400> 50  
 Met Gly Val Trp Leu Leu Ser Leu Ile Thr Val Leu Leu Ser Leu Ala  
 1 5 10 15

18

Leu Glu Thr Ala Gly Gln Gly Glu Arg Ile Ile Asp Gly Ile Lys Cys  
 20 25 30  
 Lys Glu Gly Ser His Pro Trp Gln Val Ala Leu Leu Lys Gly Asn Gln  
 35 40 45  
 Leu His Cys Gly Gly Val Leu Val Asp Lys Tyr Trp Val Leu Thr Ala  
 50 55 60  
 Ala His Cys Lys Met Gly Gln Tyr Gln Val Gln Leu Gly Ser Asp Lys  
 65 70 75 80  
 Ile Gly Asp Gln Ser Ala Gln Lys Ile Lys Ala Thr Lys Ser Phe Arg  
 85 90 95  
 His Pro Gly Tyr Ser Thr Lys Thr His Val Asn Asp Ile Met Leu Val  
 100 105 110  
 Arg Leu Asp Glu Pro Val Lys Met Ser Ser Lys Val Glu Ala Val Gln  
 115 120 125  
 Leu Pro Glu His Cys Glu Pro Pro Gly Thr Ser Cys Thr Val Ser Gly  
 130 135 140  
 Trp Gly Thr Thr Thr Ser Pro Asp Val Thr Phe Pro Ser Asp Leu Met  
 145 150 155 160  
 Cys Ser Asp Val Lys Leu Ile Ser Ser Arg Glu Cys Lys Lys Val Tyr  
 165 170 175  
 Lys Asp Leu Leu Gly Lys Thr Met Leu Cys Ala Gly Ile Pro Asp Ser  
 180 185 190  
 Lys Thr Asn Thr Cys Asn Gly Asp Ser Gly Gly Pro Leu Val Cys Asn  
 195 200 205  
 Asp Thr Leu Gln Gly Leu Ala Ser Arg Gly Thr Tyr Pro Cys Gly Gln  
 210 215 220  
 Pro Asn Asp Pro Gly Val Tyr Thr Gln Val Cys Lys Tyr Lys Arg Trp  
 225 230 235 240  
 Val Met Glu Thr Met Lys Thr His Arg  
 245

201204-021102